Neuroradiology

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History: 30 year old man with acute headache, nausea, and vomiting

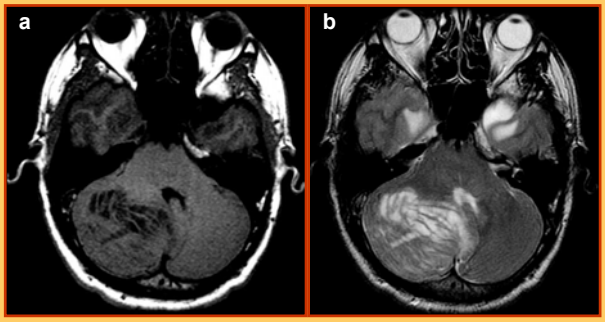

Figure 1. Axial MR a) T1W and b) T2W

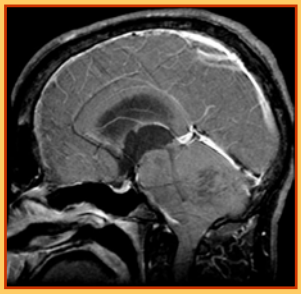

Figure 2. Sagittal T1W MR w/Gadolinium

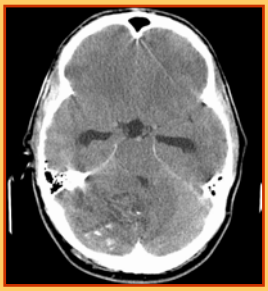

Figure3. Axial CT

Findings: On MR there is a nonenhancing mass with low T1 and high T2 signal with a laminated or layered appearance. CT shows a mass in the right cerebellum with linear areas of low attenuation and nodular calcification and mass effect on the 4th ventricle causing obstructive hydrocephalus.

Diagnosis: Lhermitte-Duclos disease

Discussion: Lhermitte-Duclos (LD) disease (dysplastic cerebellar gangliocytoma) is rare (<100 reported cases) and may be seen with Cowden phakomatosis - an autosomal dominant disorder with mucocutaneous lesions and cancer of the breast, thyroid, colon and adnexa. Patients may be aymptomatic or have intracranial hypertension, cerebellar signs are usually mild or absent. LD has been variously described as a hamartoma, low-grade neoplasm, or a developmental anomaly.

CT demonstrates a non enhancing low attenuation mass that may calcify. MR shows a non enhancing T1 low signal and T2 high signal mass in the cerebellum with a striated pattern.

Treatment is surgery, which offers good prognosis with occasional rare cases of reoccurrence.

Figure 3. LD resection, showing large dysplastic cerebellar folia (asterisks), adjacent to thinner normal folia.

References:

1. Lhermitte J, Duclos P. Sur un ganglionuroma diffus du cortex du cervelet. Bull Assoc F Etude Cancer 1920; 9:99-107.

2. Spaargaren L, Cras P, Bomhof MA, Lie ST, et al. Contrast enhancement in Lhermitte-Duclos disease of the cerebellum: correlation of imaging with

Neuroradiology

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History: **26 year old man with a prior diagnosis, a new onset of incontinence, and "cape-like" numbness over his back for 3 months.**

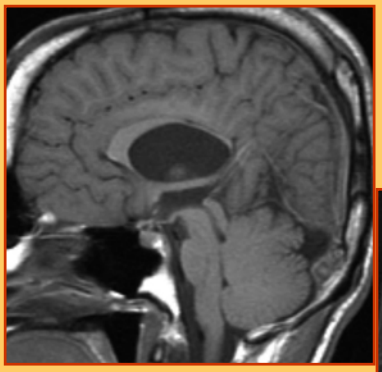

Figure 1. Sagittal T1W MR

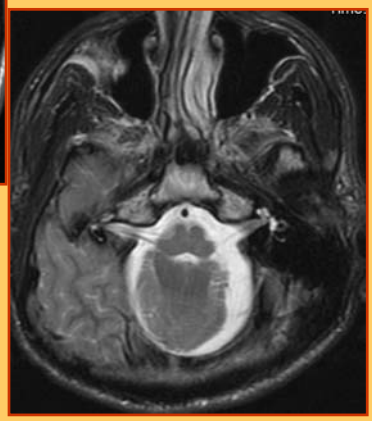

Figure 2. Axial T2W MR

Findings: **Brain MRI demonstrates agenesis of the cerebellar vermis with fusion of the cerebellar hemispheres and transversely oriented folia.**

Diagnosis: **Rhombencephalosynapsis**

Discussion: **Rhombencephalosynapsis (RS) is a rare developmental anomaly of the cerebellum, where the hemispheres are fused across the midline with absence of the vermis. Other associated anomalies include supratentorial midline defects and heterotopias. The clinical presentation varies from early death to only mild cerebellar dysfunction and developmental delay.**

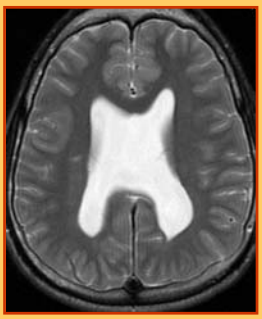

Figure 3. Axial T2W MR showing absence of the septum pellucidum and mild ventriculomegaly

References

1. Truwit CL. Barkovich AJ. Shanahan R. Maroldo TV. MR imaging of rhombencephalosynapsis: report of three cases and review of the literature. AJNR **1991**;12(5): 957-65

2. Montull C. Mercader JM. Peri J. Martinez Ferri M. Bonaventura I. Neuroradiological and clinical findings in rhombencephalosynapsis. *Neuroradiology*. **2000**;42(4): 272-4

3. Guyot LL. Kazmierczak CD. Michael DB. Adult rhombencephalosynapsis. Case report. *Journal of Neurosurgery*. **2000**;93(2): 323-5

4. Simmons G. Damiano TR. Truwit CL. MRI and clinical findings in rhombencephalosynapsis. *Journal of Computer Assisted Tomography*. **1993**;17(2): 211-4

Neuroradiology

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History: **21 month old boy with high fever, mental status changes and possible seizure.**

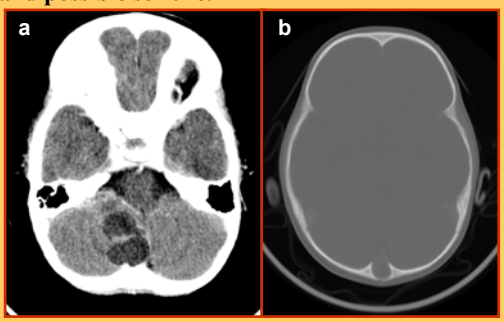

Figure 1. Axial CT a) Non-contrast and 1b) "bone window" image

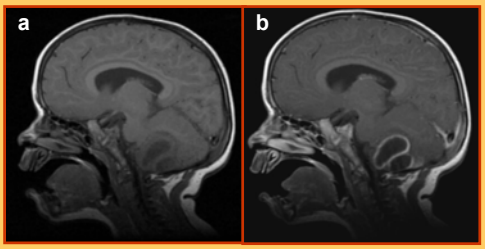

Figure 2. Sagittal T1W MR a) without and b) with Gadolinium enhancement

Findings: **Multiloculated cystic posterior fossa mass, with rim enhancement and corticated erosive remodeling of skull. The lesion is both intra-and extra-axial, and shows 'water-like' attenuation on CT.**

Diagnosis: **Infected dermoid inclusion cyst**

Discussion: **Dermoid inclusion cysts represent a failure of separation of the neural tube from the surface ectoderm. These are true cysts, lined by an epithelium, and may be associated – as in this case – with a dermal sinus tract. The lining may include these ectodermal derivatives: squamous epithelium, sebaceous glands, sweat glands, and hair follicles. Ascending infection from communication of the fistulae with the skin surface may cause meningitis – as in this case. An acute abscess would not produce the bone changes seen here. A bland dermoid may also show rim enhancement.**

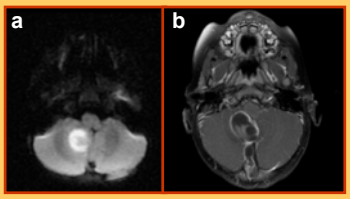

Figure 3. a) DWI shows markedly restricted diffusion, b) T1W MR after Gadolinium shows rim enhancement that extends along the sinus tract toward the skin surface.

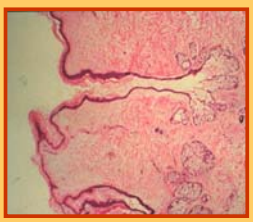

Figure 4. HE Photomicrograph showing squamous epithelium and sebaceous glands characteristic for dermoid inclusion cyst.

References:

Akhaddar, A, Mohamed J. Cerebellar abscesss secondary to occipital dermoid cyst with dermal sinus: case report. Surg Neurol 2002;58:266-70.

Neuroradiology

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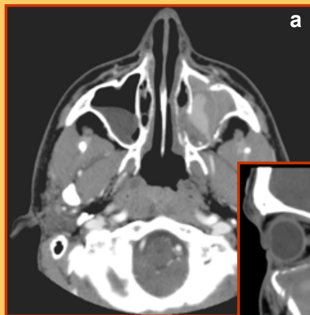
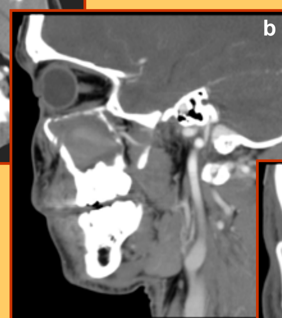
History: 22 year old man with facial trauma 2 months ago, now presents with complaint of "sinus pressure".





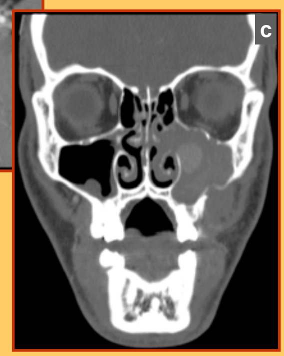
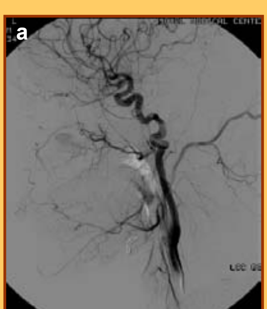


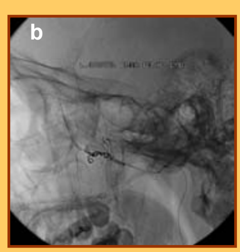
Figure1. Contrast CT a) axial,
b) sagittal, and
c) coronal

Findings: CT angiography of the face shows fractures of the left maxillary sinus, with an arterial phase contrast jet into the sinus emanating from a branch of the left internal maxillary artery. Contrast pools in the left maxillary sinus, surrounded by an older hematoma.

Diagnosis: Traumatic IMAX pseudoaneurysm

Discussion: Post-traumatic pseudoaneurysms result from partial transection of an arterial wall, secondary to either blunt force or penetrating trauma. A hematoma communicating with the vessel lumen forms and then undergoes cavitation. A fibrous pseudocapsule may develop around the hematoma, creating a pulsatile mass.





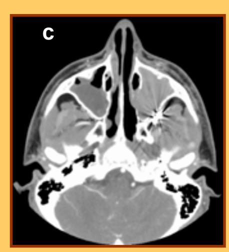


Figure 3. a) Pre and b) post embolization of IMAX pseudoaneurysm. c) Axial CT w/contrast one day post-embolization with metallic coil and no evidence of extravasation into the maxillary sinus.

References

1. D'Orta JA, Shatney CH. Post-traumatic pseudoaneurysm of the internal maxillary artery. J Trauma. 1982 Feb;22(2):161-4.
2. Conner WC 3rd, Rohrich RJ, Pollock RA. Traumatic aneurysms of the face and temple: a patient report and literature review, 1644 to 1998. Ann Plast Surg. 1998 Sep;41(3):321-6.
3. Diaz-Daza O, Arraiza FJ, Barkley JM, Whigham CJ. Endovascular Therapy of Traumatic Vascular Lesions of the Head and Neck. Cardiovasc Intervent Radiol. 2003 Apr 28 [Epub ahead of print]
4. Stehling MK, Lawrence JA, Weintraub JL, Raptopoulos V. CT angiography: expanded clinical applications. AJR Am J Roentgenol. 1994 Oct;163(4):947

Neuroradiology

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History: 59 year old woman with hearing loss on the left.

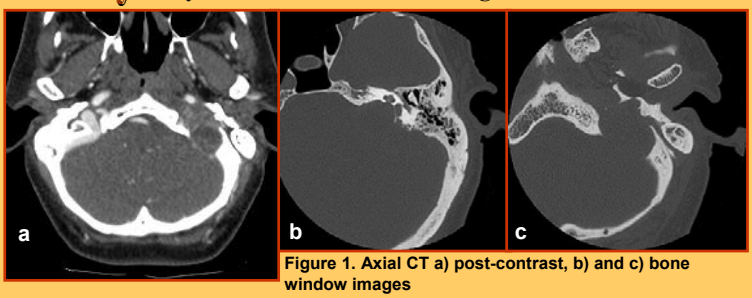

Figure 1. Axial CT a) post-contrast, b) and c) bone window images

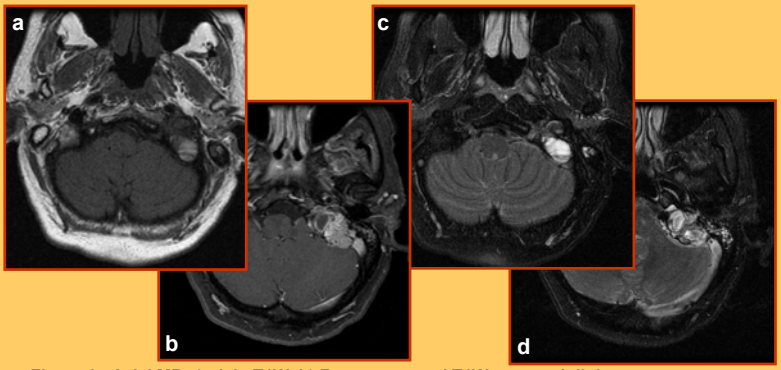

Figure 2. Axial MR a) plain T1W, b) Fat-suppressed T1W post-gadolinium; c) and d) Fat-suppressed T2 weighted FSE

Findings: CT displays a heterogeneously enhancing mass within and occluding the left jugular bulb. Precontrast T1 weighted MR shows a mixed iso- and high signal intensity mass involving the jugular foramen. Post contrast MR shows enhancement of the anterior portion of the mass within the jugular foramen. The high signal on the T1 and T2 weighted images likely represents extracellular methemoglobin. Peripherally the mass shows a rim of low signal consistent with hemosiderin. The origin of this mass, in the region of the endolymphatic duct and sac, is well demonstrated, along with involvement of the IAC and jugular bulb.

Diagnosis: Endolymphatic Sac Tumor

Discussion: Endolymphatic sac tumors (ELST) are low grade adenocarcinomas of endolymphatic sac origin (1). Slowly growing but locally aggressive neoplasms, the age range is 15 to 75 years. Symptoms include hearing loss, tinnitus, vertigo, ataxia and facial nerve palsy (2). ELST presents sporadically or with von Hippel-Lindau disease (3).

CT shows a destructive mass centered on the retrolabyrinthine petrous temporal bone, in the expected region of the endolymphatic sac and vestibular aqueduct. Extension of tumor into the posterior fossa and medial mastoid bone is common. Calcifications within the mass are often seen and may represent residual fragments of temporal bone. The mass may extend inferiorly into the jugular foramen - as in this case.

MRI reveals isointense solid areas that enhance with gadolinium. Cystic regions can be seen, with T1 shortening consistent with proteinaceous fluid and/or hemorrhage. Differential diagnosis includes paraganglioma, meningioma, squamous cell carcinoma, and primary or metastatic bone neoplasms.

References:

1. Heffner DK. Low-grade adenocarcinoma of probable endolymphatic sac origin; a clinicopathologic study of 20 cases. *Cancer* 1989;64:2292-2302